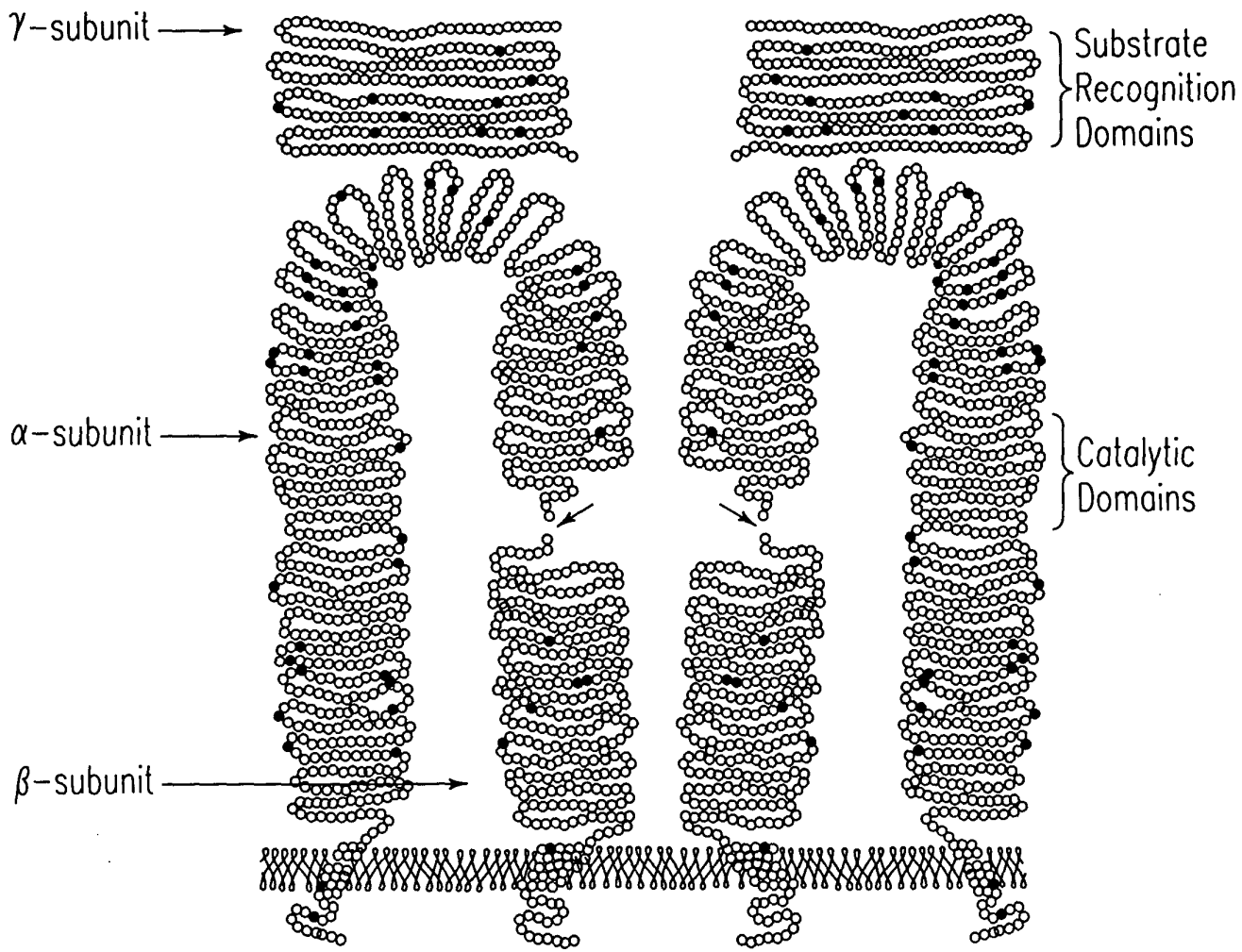
FIG. 1


γ-subunit
α-subunit
β-subunit
Substrate
Recognition
Domains
Catalytic
Domains

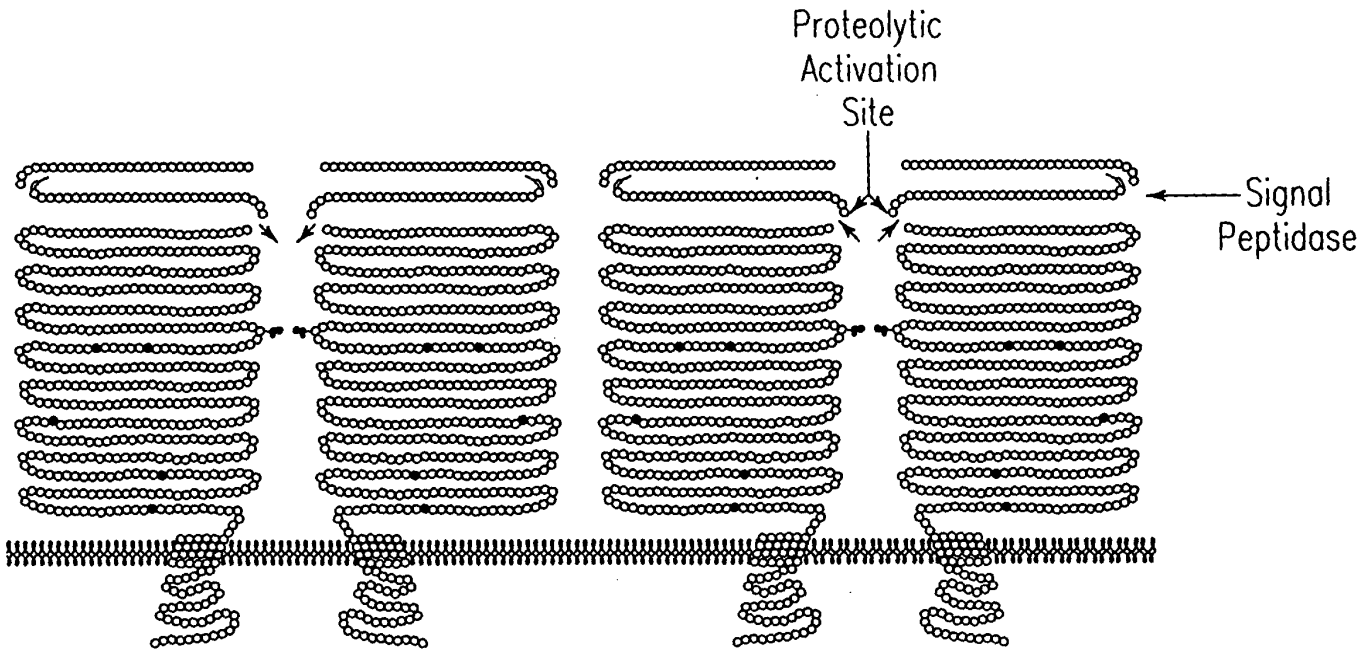
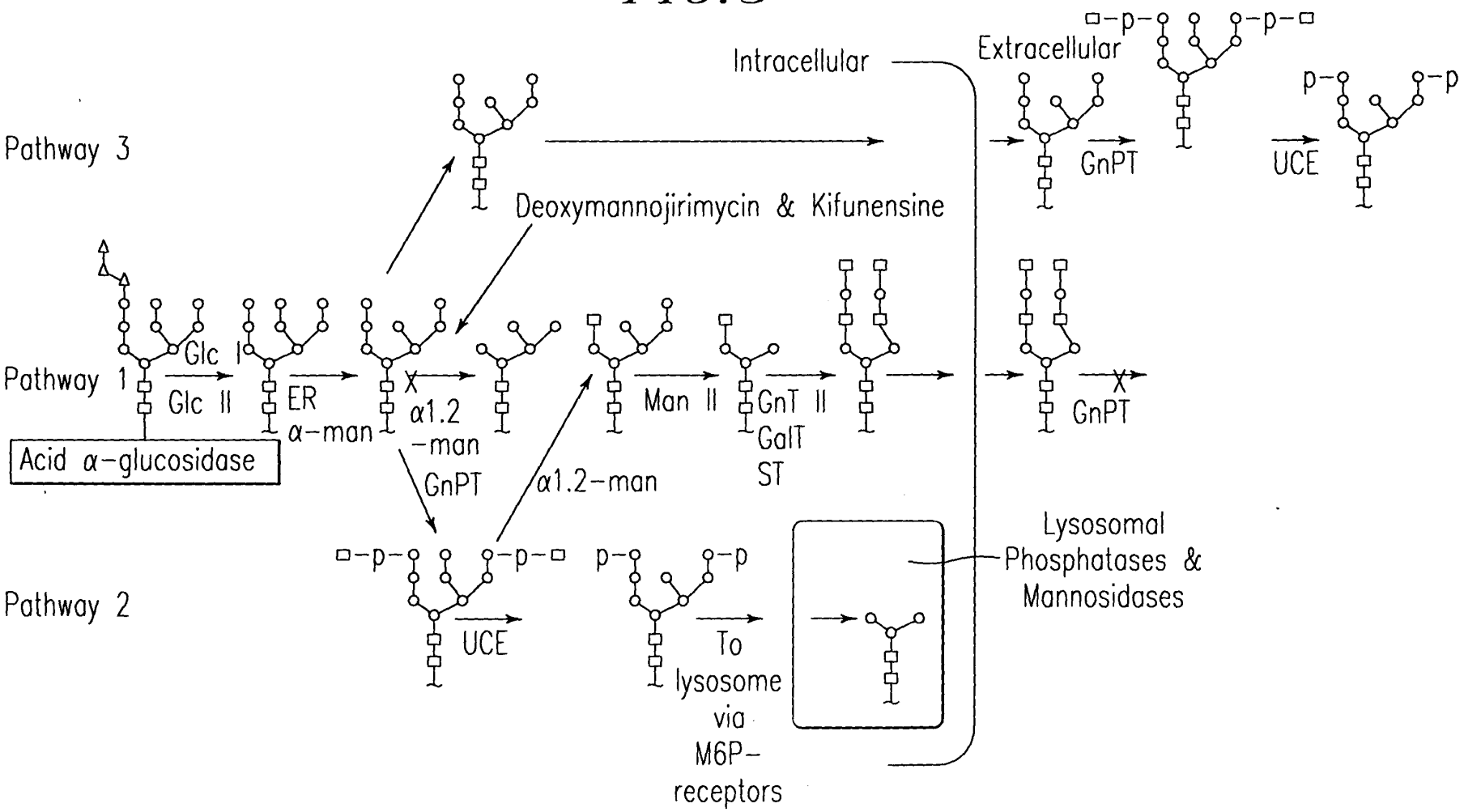


FIG. 2

FIG. 3


Intracellular
Extracellular
Pathway 3
Deoxymannojirimycin & Kifunensine
GnPT
UCE
Glc I
Glc II
ER
α-man
α1.2
-man
GnPT
Man II
GnT II
GalT
ST
GnPT
Acid α-glucosidase
Pathway 1
α1.2-man
Lysosomal
Phosphatases &
Mannosidases
Pathway 2
UCE
To
lysosome
via
M6P-
receptors

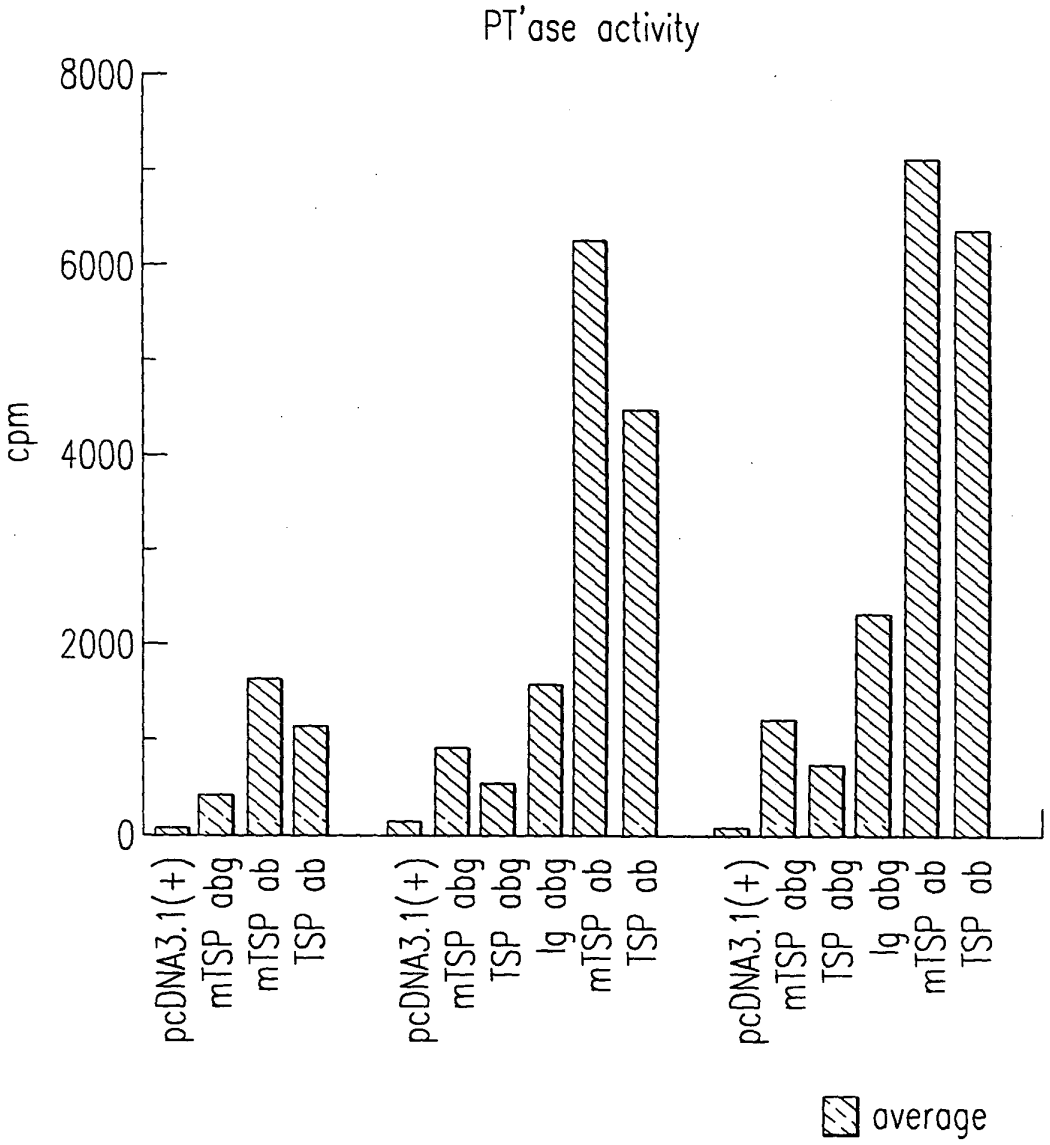

PT'ase activity
8000
6000
4000
2000
0
cpm
pcDNA3.1(+)
mTSP abg
mTSP ab
TSP ab
pcDNA3.1(+)
mTSP abg
TSP abg
Ig abg
mTSP ab
TSP ab
pcDNA3.1(+)
mTSP abg
TSP abg
Ig abg
mTSP ab
TSP ab
overage

FIG. 4